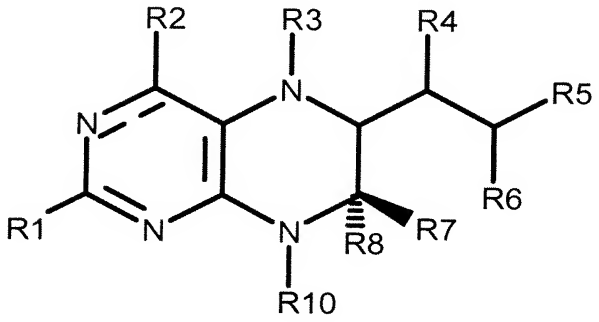
CLAIMS

1-44. Canceled

45. (previously presented) A method for long term treatment of conditions of reduced protein tolerance due to reduced phenylalanine oxidation without deficiency of cofactor tetrahydrobiopterin, said conditions caused by mutations in the phenylalanine hydroxylase gene associated with at least one of the following allele pairs:

A403V + IVS4+5G>T, P314S + R408W, F39L + D415N, Y414C + D415N, Y417H + Y417H, F55L + S310Y, V177M + R408W, P275L + Y414C, V245A + R408W, L48S + R158Q, Y417H + Y417H, V245A + R408W, R261X + A300S, R158Q + E390G, Y414C + IVS12+1G>A, I65S + A300S, H170O + A300S, R261Q + Y414C, K274fsdel11bp + E390G, IVS4-5C>G + R408W, I65T + Y414C, E390G + IVS12+1G>A, I65V + R261Q, R158Q + Y414C.

said method comprising administering a medicament containing at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-Acyl, wherein the Acyl residue contains 1 to 32 carbon atoms;

wherein R2 is selected from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, Acetyl, OX, wherein X is a C1 to C32 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 is CH_3, C_2H_5, C_3H_7, or butyl;

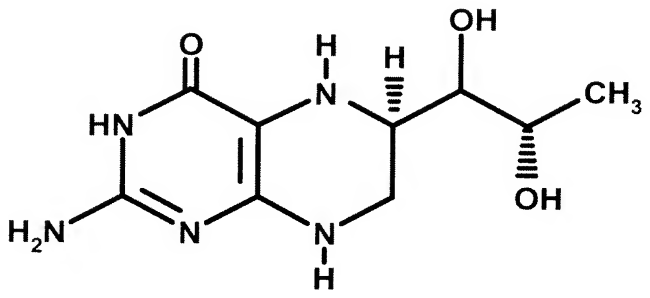
wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and - - represents an optional double bond;

as well as their pharmaceutically acceptable salts.

46. (previously presented) A method as in claim 45, wherein said medicament is administered to a patient in need thereof until said patient exhibits improvement in protein tolerance.

47. (previously presented) A method as in claim 45, wherein R1 is NH-acyl, wherein the acyl residue contains CH_3O or 9 to 32 carbon atoms, and wherein at least one of R4 and R6 are C9 to C32 acyl residues.

48. (previously presented) A method according to claim 45, wherein the compound is selected from the group consisting of: 5,6,7,8-tetrahydrobiopterin, sapropterin, a compound having the following structure:



(-)-(1'R,2'S,6R)-2-Amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterin; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterin; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterin; and/or

2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterin.

49. (previously presented) A method according to claim 45, wherein said pharmaceutically acceptable salt a hydrochloride or a sulphate.

50. (previously presented) A method according to claim 45, wherein the condition of reduced protein tolerance is at least one of: conditions with increased phenylalanine or reduced tyrosine in body fluids, tissues or cells.

51. (previously presented) A method as in claim 50, wherein said condition of reduced protein tolerance is classical phenylketonurea, mild phenylketonurea or mild hyperphenylalaninemia

52. (previously presented) A method according to claim 45, wherein said medicament functions as chaperone for improving protein folding, in particular in the case of structural anomalies of enzymes, which require tetrahydrobiopterine as a cofactor.

53. (previously presented) A method according to claim 52, wherein said enzyme is selected from phenylalanine hydroxylase, tyrosine hydroxylase, tryptophane hydroxylase, or NO-synthase.

54. (previously presented) A method according to claim 45, wherein said compound functions as chaperon as neurotransmitter and/or second messenger enhancer, in particular in conditions with increased phenylalanine or lowered tyrosine, serotonin or dopamine in body fluids, tissues, or cells, in particular in conditions with reduced

phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase, or NO-synthase activity.

55. (previously presented) A method according to claim 45, wherein said compound functions as neurotransmitter or as second messenger enhancer, in particular for catecholamine and/or serotonin and/or dopamine and/or nitrogen oxide (NO).

56-62. Canceled.